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Title: Hemodynamics and response to therapy of pulmonary hypertension in patients with interstitial lung disease: Preliminary results of the “HYPID” prospective study

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Body: Background. Pulmonary hypertension (PH) is associated with a shorter survival in patients with interstitial lung disease (ILD). Objective. To study the characteristics and response to specific PH therapy in patients with ILD and precapillary PH at right heart catheterization. Methods. A prospective multicenter observational study was conducted in French expert centers for rare pulmonary diseases or PH (NCT01443598). Results. The first 100 patients were studied (mean age 64 ± 10 years; 63 males). ILD was idiopathic pulmonary fibrosis (n=25), combined pulmonary fibrosis and emphysema syndrome (n=21), systemic sclerosis with ILD (n=26), sarcoidosis (n=8) and other ILD (n=20). Overall, NYHA class was I-II in 24% of patients and III-IV in 76%. Six-min walk distance was 289 ± 140m. Hemodynamic characteristics were mPAP 38 ± 9.8 mmHg, CI 2.7 ± 0.65 L/min/m², and PVR 515 ± 243 dyn.s.cm-5. mPAP was >35 mmgHg (“disproportionate” PH) in 56% of the cases. No correlation was found between pulmonary function and hemodynamic parameters. No differences were found between the 4 etiological groups for hemodynamic parameters. 36 patients with mPAP >35 mmHg received PH therapy and were evaluated at 3-6 months; mPAP decreased from 46 to 38 mmHg ($p<0.0001$), CI improved from 2.6 to 2.9 L/min/m² ($p<0.05$), and PVR decreased from 640 to 460 dyn.s.cm-5 ($p<0.0001$), but no difference was observed in 6

min walk distance or NYHA class. Conclusion. Hemodynamic characteristics do not correlate with pulmonary function and are comparable between etiological categories of ILD with PH. Preliminary data suggest that PH therapy may improve hemodynamics.